ORIGINAL ARTICLE

Hirchsprung Disease in Adults & Adolescence

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ABSTRACT

Background: Hirchsprung disease or aganglionic colon is a rare disease in children but could be manifested undiagnosed I adults in later life.
Aim: To diagnose and treat adults and adolsencents for missed Hirchsprung disease
Duration and place of study: Five patients with Hirchsprung disease were treated in Ittefaq hospital Trust, Lahore from August 2015 until February 2016
Results: All treated patients who were adults did extremely well with the aganglionic megacolon Diagnosis and treatment provided successfully with the hospital resources.
Conclusion: Good in good surgical hands.
Keywords: Hirchprung disease, adolescence

INTRODUCTION

Hirschsprung disease is intestinal disease due to innervation disorders. These disorders are known as intestinal neuronal dysplasia. Histopathologically it consists of aganglionosis of the rectum or colon and manifests during infancy or childhood. However, milder forms of the disease may go undiagnosed and progress to adulthood, Colonic region proximal to the distal obstructed segment dilates due to a compensatory role. Patients can manage this condition by using cathartic agents, bulk formers, and enemas. Ultimately, the dilated proximal colonic segment may decompensate secondary to distal physiologic obstruction, and patients may experience rapidly worsening constipation or acute obstruction. This disease can be overlooked and misdiagnosed in adults. Therefore, adult HD is thought to be more common than previously recognized. HG, which can mimic HD, a reduced number of parasympathetic nerves in the intestinal wall.

Adult patients with HD or HG have the similar clinical manifestation of a lifelong history of refractory constipation. However, these patients often have severe symptoms or complications with erosion and ulceration, which cause bleeding or perforation of the colon, partial or complete intestinal obstruction.

Therefore, an accurate diagnosis of adult HD is essential, given the curability of these disorders with surgery. Also, the differentiation between adult HD and adult HG is helpful in formulating a preoperative plan because the disease extent of adult HD is mainly limited to the rectosigmoid colon and requires localized surgery.

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Diagnosis: The diagnosis of HD or HG is usually much more difficult in adults than in infants, with the final diagnosis based on the results of full-thickness biopsy of the intestinal wall. However, imaging studies such as computed tomography (CT) and double-contrast barium enema examination are generally accepted procedures for evaluating chronic constipation, which is a common disorder in adults.

MATERIALS AND METHODS

Yusra Begum 20 years old married woman from Kasur and 4 other patients age 18 to 22 were seen with chronic constipation , weight loss and miscarriage .They were investigated by colonoscopy and double contrast Barium enema at our institution between August 2015 and December 2005 in Ittefaq Hospital, Trust Lahore Pakistan.

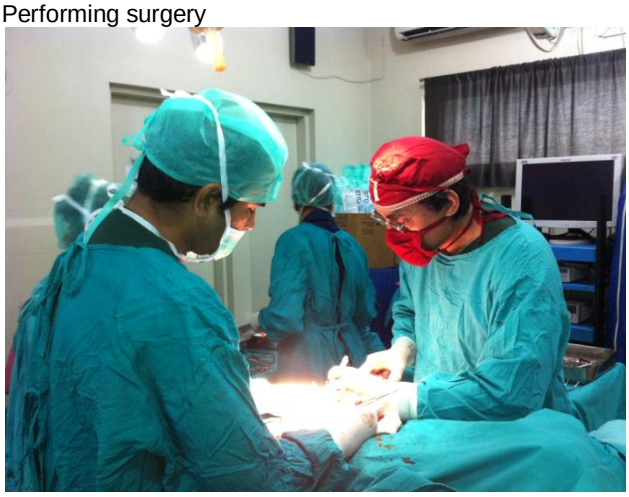
Data collection: Yusra and others had constipation since childhood relieved by lexatives and enemas. Yusra had miscarriage a year ago while 3 months pregnant. The diagnoses of all patients were histopathologically confirmed with full-thickness rectal biopsy, and all 5 patients underwent surgery with low anterior resection and covering ileostolies, rectosigmoidectomies and pull through operation which were successfully reversed from 3 to 6 months . The outcomes of these patients after surgery, including any change in bowel movement or body weight, were also recorded.

CT and Barium Enema Examination and Colonoscopy: CT scans had been obtained in all patients by using one of three with colonocopy and deep biopsies before surgical intervention carried out. Double-contrast barium enema examinations were performed in all the five patients.

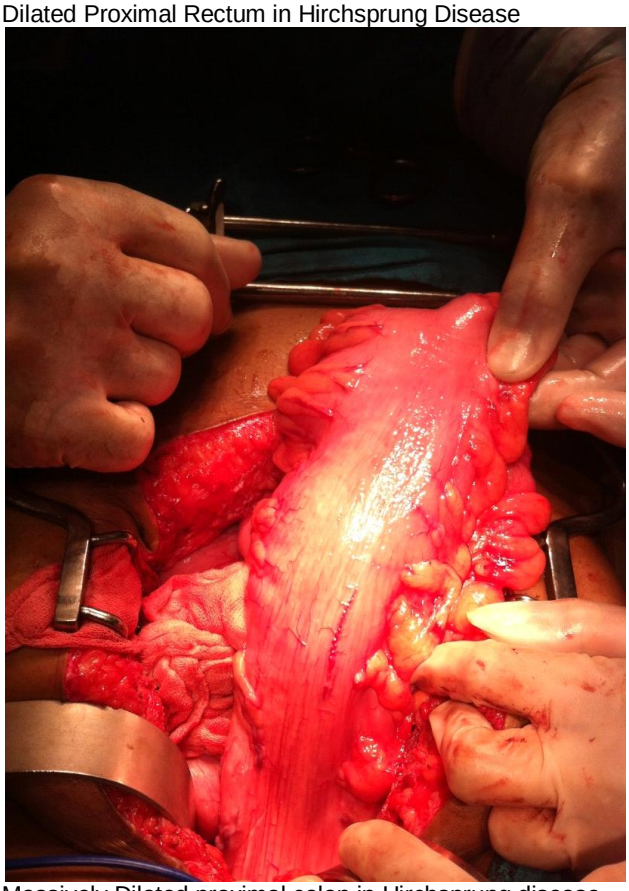
Surgical procedure: All the 5 patients underwent various procedures including laparotomy, low anterior resection recto sigmoidectomy and pull through operation and covering ileostomies performed which

were reversed between period 3 to 6 months without any problem. Histology finding of the resected specimen confirmed a ganglionic colon confirming Hichsprung disease. Routine follow up in 1 month for first 3 months and then 6 months' time proved good results .Two patients had mild diarrhea initially which settles subsequently .

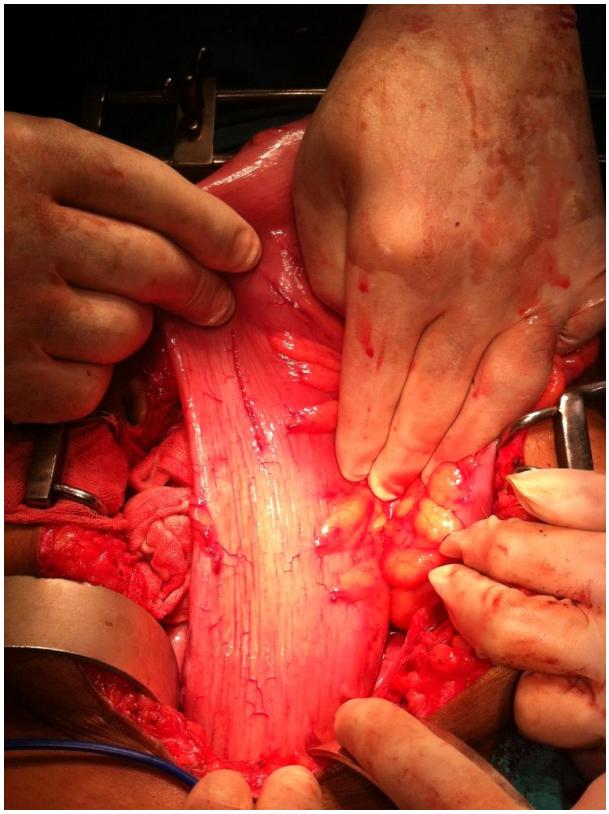
Performing surgery



Dilated Proximal Rectum in Hirchsprung Disease



Massively Dilated proximal colon in Hirchsprung disease



RESULTS

All patients had refractory chronic constipation with abdominal pain and distention. In the four patients with HD, chronic constipation had existed from infancy or childhood. In the five patients with HG, constipation was discovered during infancy or childhood. The frequency of bowel movements in these patients was once every 3–10 days for the patients with HD, once every 5–10 days for the patients with HG, and once every 4 days for the patient with combined HD and HG.

Regular use of enemas or laxatives was necessary to induce defecation in the patients .The two patients with HG that manifested during infancy or childhood also needed to use enemas or laxatives. In the two patients with HG or combined HD rectosigmoidectomy, and in other two low anterior resection and retrorectal pull-through in one patient each—according to the extent of the involved colorectal segment. All these 5 patients had satisfactory clinical outcomes with improved bowel movements and weight gain after surgical treatment.

Histopathology: In the all 3 patients with HD, an aganglionic segment was detected in the upper part of the rectum. In the 2 patients with HG, a decreased number of ganglion cells, characteristic of HG, was detected in the sigmoid colon and lower rectum.

Pathophysiology: HD occurs in approximately one in 5000 live births . Ninety-percent of these cases are diagnosed before the patient in childhood . In rare milder cases of these disorders, the patient may not receive a diagnosis until he or she reaches adulthood (20 to 23). The primary defect in adult HD is identical to that seen in infancy or childhood and is characterized by the total absence of intramural ganglion cells of the submucosal (Meissner) and myenteric (Auerbach) plexuses in the affected segment of the bowel. The aganglionic segment is the result of failure of ganglion cell precursors to migrate from the neural crest into the hindgut. The cause of adult HG is still unknown, but a previous report suggests that an inborn hypoplasia of the parasympathetic myenteric plexus is responsible. However, in several other reports, it is assumed that HG is an acquired rather than congenital disease.
Circulatory disturbances, intramural inflammation, and infectious disease have been reported as causes of adult HG. HG has been reported to have variable forms—specifically, isolated HG, localized HG, disseminated HG, and HG with intestinal neuronal dysplasia. In cases of disseminated HG, abnormal involvement of the entire colon necessitates total colectomy, whereas in adult HD, the disease is limited primarily to the sigmoid colon or the rectum, and, therefore, only localized partial colectomy or anorectal surgery is required. The correct diagnosis of HD or HG is based on the proper medical history, the barium enema examination results, and, most important, the full-thickness rectal biopsy findings.

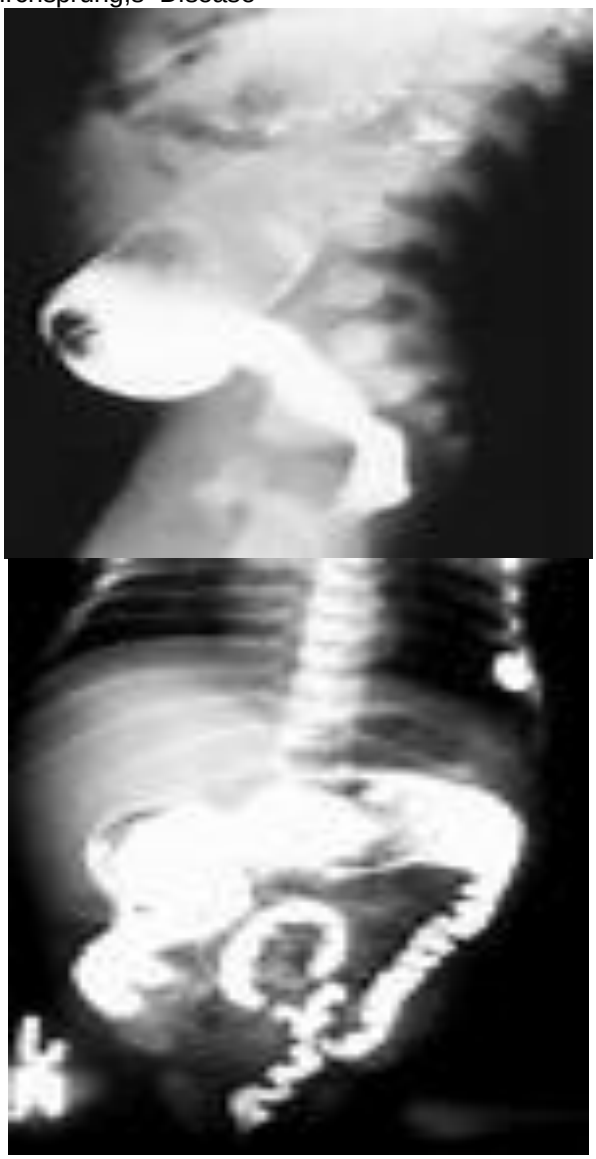
Radiographs of the abdomen usually show massive distention of the proximal region of the colon, with a small narrowed distal segment. However, rectal narrowing may not be detected in approximately 20% of patients.

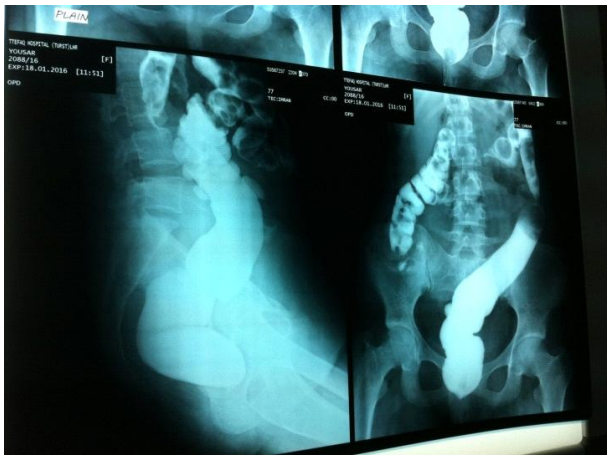
Although CT is considered a common imaging modality for excluding other diseases such as colorectal cancer, which also causes chronic constipation in adults, to our knowledge, there have been no reports regarding the CT findings of adult HD and adult HG in the radiology literature. In the cases of HD, we presumed that the proximal colonic segment was more dilated because the aganglionic segments showing HD were contracted compared with the rectums showing HG, which had scarce but extant ganglion cells.

DISCUSSION

Hirchsprung disease is usually an intractable disease of the children which can be missed until adulthood .More common in people with low socioeconomic back ground. It could be devastating by chronic constipation n, abdominal distension and miscarriage due to pressure on the uterus. It can become a crippling disease due to continuous use of laxatives, pain and distension. Usually goes undiagnosed in poor socioeconomic back ground and uneducated families and loss of awareness and facilities to reach the diagnosis. Major surgical intervention is required including bowel resection and anastomosis, anterior resection, pull through and temporary or permanent colostomy or ileostomy .Some patients had poor compliance of the reversal ,others had complications related to surgery and stomas increasing morbidity and mortality.
Surgery should be carried out in specialist colorectal unit performing surgery for this crippling disease more often.

Hirchsprung,s Disease





CONCLUSION

On imaging a markedly dilated feces-filled proximal colonic segment with a transition zone and a narrowed distal colonic segment without any other obstructive cause depicted on CT and double-contrast barium enema images, in conjunction with a clinical history of chronic refractory constipation, in an adult should suggest a diagnosis of adult HD or adult HG. The identification of a much higher transition zone ratio may help in the differential diagnosis of adult HD versus adult HG and in the guidance of treatment for these diseases.

Conflict of interest: Author has no conflict of interest with anyone.

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